# LONG TERM OUTCOME IN Wilms' tumor

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# INTRODUCTION

Nephroblastoma, or Wilms' tumor, is an embryonal tumor that develops from remnants of immature kidney. It is the most common renal tumour of childhood.

## NWTS-5 protocol

Stage I: The tumor is limited to the kidney and was completely excised. The renal capsule has an intact outer surface. The tumor was not ruptured or biopsied prior to removal (fine-needle aspiration biopsies are excluded from this restriction). The vessels of the renal sinus are not involved. There is no evidence of tumor at or beyond the margins of resection.

Stage II: The tumor extends beyond the kidney, but was completely excised. There may be regional extension of tumor (i.e. penetration of the renal capsule or extensive invasion of the renal sinus). The blood vessels outside the renal parenchyma, including those of the renal sinus, may contain tumor. The tumor was biopsied (except for fine-needle aspiration), or there was spillage of tumor before or during surgery that is confined to the flank, and does not involve the peritoneal surface. There must be no evidence of tumor at or beyond the margins of resection.

Stage III: Residual non-hematogenous tumor is present, and confined to the abdomen. Any one of the following may occur:

(1) Lymph nodes within the abdomen or pelvis are found to be involved by tumor (renal hilar, para-aortic, or beyond). (Lymph node involvement in the thorax, or other extra-abdominal sites would be a criterion for stage IV.)

(2) The tumor has penetrated through the peritoneal surface.

(3) Tumor implants are found on the peritoneal surface.

(4) Gross or microscopic tumor remains postoperatively (e.g. tumor cells are found at the margin of surgical resection on microscopic examination). (5) The tumor is not completely resectable because of local infiltration into vital structures. (6) Tumor spill not confined to the flank occurred either before or during surgery.

Stage IV: Hematogenous metastases (lung, liver, bone, brain, etc.), or lymph node metastases outside the abdominopelvic region are present.

Stage V: Bilateral renal involvement is present at diagnosis. An attempt should be made to stage each side according to the above criteria on the basis of the extent of disease prior to biopsy or treatment.

# AIM OF THE STUDY

- Our aim is to study the long term outcome in 156 children with Wilms' tumor from december1999 - december 2009 in ICH .

- To analyze long term outcome in wilms tumour in perplex situations as double moiety, wilms with atrial thrombi, wilms in syndromic variety & bilateral Wilms', wilms in horseshoe kidney and extrarenal wilms

- Our long term follow-up included time of menarche, skeletal growth, hyper tension , cardio toxicity due to usage of adriamycin & effects of radiotherapy.

- To analyze the time of resolution of IVC thrombus and effect of neo adjuvant therapy in horseshoe kidney, solitary kidney, bilateral wilms tumour, large tumour ,distant metastasis.

# REVIEW OF LITERATURE

National Wilms' Tumor Study Group

The first two NWTSG studies, NWTS-1(1969–1973) and NWTS-2 (1974–1978), showed that postoperative local irradiation was unnecessary for group I patients.[96,97] The combination of vincristine (VCR) and dactinomycin (AMD) was noted to be more effective than the use of either drug alone, and the addition of doxorubicin (DOX) improved survival for higher-stage patients. Important findings of NWTS-1 and -2 were identification of unfavourable histologic features and other prognostic factors that allowed refinement of the staging system, stratifying patients into high-risk and low-risk treatment groups.[18] It was recognized that the presence of lymph node metastases had an adverse outcome on survival.[16,17] Children with lymph node metastases as well as those with diffuse tumor spill were found to be at increased risk of abdominal relapse. Therefore, such patients were from then on classified as having stage III disease and given whole abdominal irradiation. These findings were incorporated into design of NWTS -3 to try and decrease the intensity of therapy for majority of low risk patients.

In NWTS-3 (1979–1986), patients with stage I, FH Wilms' tumor were treated successfully with either a 10- or 18-week regimen of VCR

and AMD. This considerably decreased the amount of chemotherapy administered and the total duration of treatment. The 4-year relapse-free survival was 89%, and the overall survival was 95.6%. Stage II FH patients treated with AMD and VCR without postoperative XRT had an equivalent survival (4 - year overall survival of 91.1%) to patients who received the same treatment plus DOX with or without XRT. This demonstrated that the cardiotoxic drug DOX is not necessary for the successful treatment of this group of patients. This also demonstrated that XRT could now be omitted for the majority of childrenwith Wilms' tumor. for stage III FH patients, 10.8Gy of abdominal irradiation was shown to be as effective as 20 Gy in preventing abdominal relapse if DOX was added to VCR and AMD. The 4-year relapse-free survival for stage III patients was 82% in NWTS-3 and the 4-year overall survival was 90.9%.

Patients with stage IV FH tumors received abdominal (local) irradiation based on the local tumor stage. In addition, they all received 12 Gy to both lungs. In combination with VCR, AMD, and DOX, the 4-year relapse-free survival was 79% and the overall survival was 80.9%. There was no statistically significant improvement in survival when cyclophosphamide was added to the three-drug regimen.

The goals of NWTS-4 (1986–1994) were to continue improving treatment results while decreasing the cost of therapy through modification of the schedule of drug administration. ‘Pulse-intensive’ chemotherapy regimens, employing single doses of AMDand DOX, were compared with regimens using divided doses of the drugs. Pulse-intensive regimens utilized simultaneous administration of agents at less frequent intervals to decrease the number of clinic visits and hence the cost of cancer treatment. In addition, treatment durations of approximately 6 and 15 months were compared in patients with stages II–IVFH tumors. NWTS-4 demonstrated that, while the administered drug dose–intensity was greater on pulse-intensive regimens, these regimens produce less hematologic toxicity than the standard regimens.19 Patients treated with pulse-intensive regimens achieved equivalent survival compared with those treated with standard chemotherapy regimens.20Treatment with 6 months of chemotherapy was as effective as 15 months.

Children with anaplastic Wilms' tumors were randomized in NWTS-3 and NWTS-4 to receive VCR,AMD, DOX, or those three drugs with the addition of cyclophosphamide. The results were analyzed after the tumors were reclassified using the criteria of Fariaet al.63 There was no difference in outcome between the regimens for children with focal anaplasia, who had a prognosis similar to that for favorable histology patients.21 For stage II–IV diffuse anaplasia, the addition of cyclophosphamide to the three-drug regimen improved the 4-year relapse-free survival (27.2%vs 54.8%).

Treatment of patients with stage I or II FH tumors, and stage I anaplastic Wilms' tumor was the same. They received a pulse-intensive regimen of VCR and AMD for 18weeks. Patients with stage III FH and stage II–III focal anaplasia were treated with AMD, VCR, and DOX, and 10.8 Gy XRT. Patients with stage IV FH tumors received abdominal irradiation based on the local tumor stage and 12 Gy to both lungs. NWTS-5 was a single-arm therapeutic trial without any randomization for therapy. Prospective collection of information regarding biologic features of the tumors was part of this study. A collection of banked tumor specimens is available to evaluate new prognostic factors that may be identified in the future.

Most importantly, the clinical outcome is available for patients for whom there are banked specimens. One of the primary goals of the study was to verify the preliminary findings that loss of heterozygosity (LOH) for chromosomes 16q and 1p is useful in identifying patients at increased risk for relapse and death. Among patients with stage I–II FH tumors, the relative risk (RR) of relapse and death were increased for LOH 1p only (RR = 2.2 for relapse; RR = 4.0 for death), for LOH 16q only (RR = 1.9 and RR = 1.4),and for LOH for both regions (RR = 2.9 and RR = 4.3) in comparison with patients lacking LOH at either locus.52 The risks of relapse and death for patients with stage III–IV FH tumors were increased only with LOH for both regions (RR = 2.4 and RR = 2.7). These results demonstrate that LOH for these chromosomal regions can now be used as an independent prognostic factor,

**Treatment scheme used in wilms tumour study-5**

| | Radiotherapy | Chemotherapy |
|---|---|---|
| Stage I and II, favorable histology | None | Regimen 4A |
| Stage I focal or diffuse anaplasia | | |
| Stage I, favorable histology, age <2 years, tumor weight <550 g | None | Regimen EE-4A |
| Stage III and IV, favorable histology | yes | Regimen DD-4A |
| Stage II–IV, focal anaplasia | | |
| Stage II–IV, diffuse anaplasia | Yes | Regimen I |
| Stage I–IV clear cell sarcoma of the kidney | | |
| Stage I–IV rhabdoid tumor of the kidney | Yes | Regimen RTK |

a 1998 modification to original protocol:

- Regimen EE-4A: pulse-intensive dactinomycin, vincristine (18 weeks)
- Regimen DD-4A: pulse-intensive dactinomycin, vincristine, doxorubicin (24 weeks)

- Regimen I: dactinomycin, vincristine, doxorubicin, cyclophosphamide, and etoposide (24 weeks)

- Regimen RTK: carboplatin, etoposide and cyclophosphamide (24 weeks)

- Stage IV/FH patients are given radiation based on the local tumor stage classified as having stage III disease and given whole abdominal irradiation. These findings were incorporated into the design of NWTS-3 to try and decrease the intensity of therapy for the majority of low-risk patients.

**NWTS-5 FLOWSHEETS**: complete a column below for each day drugs are given or toxicities occur. Flowsheets and other clinical records must be submitted following all relapses.

Name: _____ Institution: _____ Page #: ____ Patient ID#: ☐☐☐☐☐☐

| | | | | | | | | |
|---|---|---|---|---|---|---|---|---|
| 1. Date | | | | | | | | |
| 2. Hospitalizations | | | | | | | | |
| 3. Height (cm) | | | | | | | | |
| 4. Weight(kg) | | | | | | | | |
| 5. $M^2$ | | | | | | | | |
| 6. Radiation/surgery | | | | | | | | |
| 7. Antibiotics | | | | | | | | |
| 8. Transfusions | | | | | | | | |
| 9. Hgb (grams) | | | | | | | | |
| 10. Platelets (x $10^3$) | | | | | | | | |
| 11. WBC (x $10^3$) | | | | | | | | |
| 12. ANC/%Neutr. | | | | | | | | |
| PHYSICAL EXAMINATION/CHEMISTRIES: | | | | | | | | |
| 13. Temp (F or C) | | | | | | | | |
| 14. B/P | | | | | | | | |
| 15. Masses/other | | | | | | | | |
| 16. SGOT/SGPT | | | | | | | | |
| 17. Bilirubin/Alk Phos | | | | | | | | |
| 18. BUN/Creatinine | | | | | | | | |
| 19. Urine protein-cells | | | | | | | | |
| IMAGING STUDIES: In REMARKS, record all results determining disease status (POS/NEG). | | | | | | | | |
| 20. X-ray | | | | | | | | |
| 21. Ultrasound | | | | | | | | |
| 22. CT Chest | | | | | | | | |
| 23. CT Abdomen | | | | | | | | |
| 24. Echo/MUGA | | | | | | | | |
| 25. Other | | | | | | | | |
| TOXICITIES: For primary therapy record toxicities and associated treatment on the roadmaps. Following primary therapy, please identify toxicities below and in more detail in the REMARKS section. | | | | | | | | |
| 26. Toxicity (specify) | | | | | | | | |
| 27. Toxicity (specify) | | | | | | | | |
| 28. Toxicity (specify) | | | | | | | | |
| INITIALS: | | | | | | | | |

**REMARKS**:



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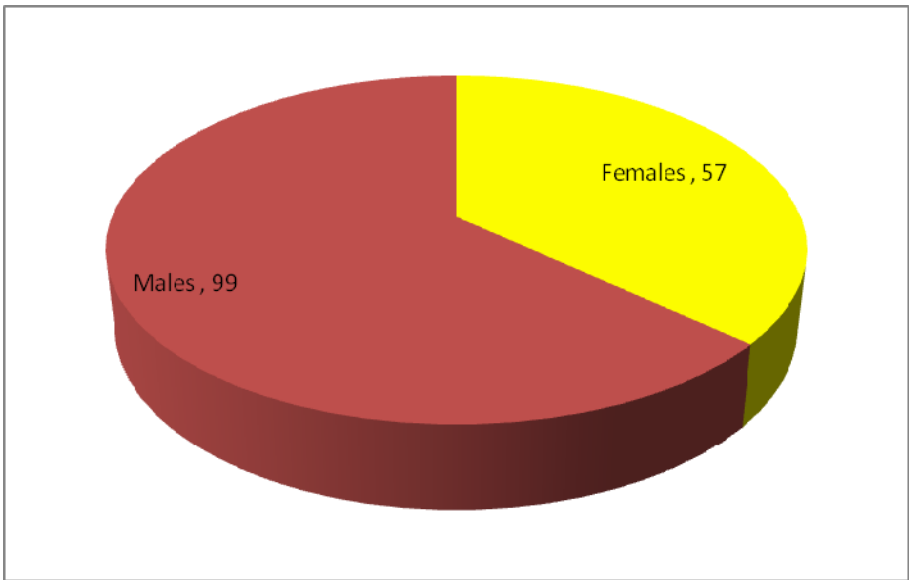
**HYPERFILTRATION damage: theory and practice**

**After nephrectomy,it is the normal response of the remaining kidney to hypertrophy.the stimulus for this hypertrophic response follows a rapid increase in blood flow in response to contralateral nephrectomy.** ***Hostetter et al drew attention to the observation that hyperfiltration in remnant nephron was a potentially adverse response to renal ablation.Rats with subtotal renal ablation developed glomerulosclerosis and progressive renal damage.***

***Rats are to more prone to develop proteinuria and reductions in glomerular filtration.***

**Rate if uninephrectomy was done in infancy versus adulthood.**

## DEMOGRAPHICS OF WILMS IN ICH-CHENNAI




**Number of wilms patients between 1999 -2009 :156**

Gender-slight male predominance

Male—99 female-57

Age at diagnosis (years)

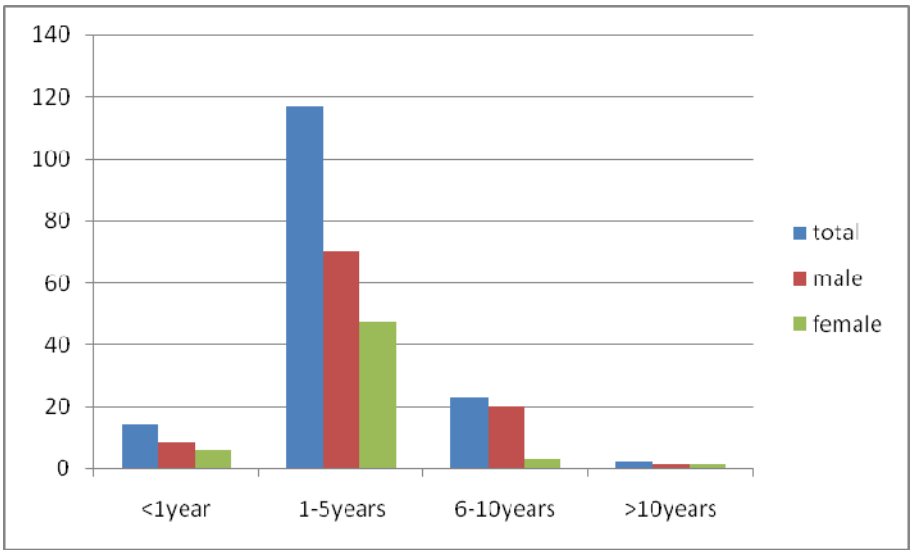
Most of the children were between 1-5 years

Number of children under 1year- 14 (8m,6f)

Number of children between 1-5- 117(70m,47f)

Number of children between 6-10-23(20m,3f)

Number of children more than10years-2(1m,1f) 10years




Initial clinical presentation

| | |
|---|---|
| Palpable abdominal mass | 137(91.2%) |
| Gross hematuria | 4 |
| Abdominal pain | 8 |
| Fever | 6 |
| Nausea/vomiting | 1 |
| Ascites | 1 |

| Associated anomaly | |
|---|---|
| Beckwith-wiedemann syndrome | 2 |
| duplex kidney | 2 |
| Congenital blindness | 1 |
| WAGR | 1 |
| Horseshoe kidney | 2 |
| Hypospadias | 1 |
| Extra-renal wilms | 1 |

## PATHOPHYSIOLOGY

More than one-third of kidneys resected for Wilms'tumor contain precursor lesions, known as nephrogenic rests.[68] Nephrogenic rests represent the abnormal persistence into postnatal life of embryonal cellsthat can produce a malignancy. Two distinct categories of nephrogenic rests exist, based on the position of these lesions within the renal lobe. Perilobar nephrogenic rests (PLNRs) are confined to the periphery of the renal lobe. Intralobar nephrogenic rests (ILNRs) occur anywhere in the kidney, including the renal sinus and collecting system. There are biologic differences distinguishing PLNR fromILNR (Table). Nephrogenic rests have a varied life and most do not form Wilms' tumor. A rest can undergo maturation, sclerosis, involution, and complete disappearance. PLNRs have been detected in1% of kidneys in infants on postmortem examination; most of these rests apparently undergo involution.[69] ILNR can become cystic and be indistinguishable from renal dysplasia. Hyperplastic nephrogenic rests can produce a renal mass easily mistaken for a small Wilms' tumor. Biopsy of a hyperplastic rest is of little value in distinguishing this lesion from a Wilms' tumor unless the interface between the rest and normal kidney of included. Wilms' tumor will have a pseudocapsule at the interface, with the normal parenchyma compressing the normal elements. Neoplastic induction of cells of a

nephrogenic rest can produce Wilms' tumor and possibly other benign or malignant renal neoplasms.**The Wilms' tumor will have a spherical shape, whereas hyperplasic rests will be more elliptical orlenticular in shape. Magnetic** resonance imaging (MRI) may be of some value in distinguishing between the two lesions, but this needs to be confirmed prospectively in large numbers of patients

| | ILNR | PLNR |
|---|---|---|
| Associated Syndromes | WAGR | BWS |
| | Hemihypertrophy | Denys–Drash |
| Genitourinary anomalies | perlman | |
| Median age of | 23 months | 36 months |

WAGR = Wilms' tumor, aniridia, genitourinary,

and retardation; BWS = Beckwith–Wiedemann

syndrome; ILNR = intralobar nephrogenic rest;

PLNR = perilobar nephrogenic rest.

Abnormal collections of blastemal cells are found in about 1%of kidneys on postmortem examination, yet these are found in approximately 40% of kidneys removed for Wilms' tumor .These histologic abnormalities are found uniformly in the kidneys of children

with an inherited susceptibility to Wilms' tumor. When multiple nephrogenic rests are found, or when the presence of rests can be inferred, such as in multicentric or bilateral Wilms' tumor, the condition is described as nephroblastomatosis. Not all nephroblastomatoses develop into Wilms' tumor; most regress or develop into a clinically insignificant benign entity. Detection of these rests is challenging on conventional imaging, and they are found only on extensive histologic search. Depending on their location in the kidney, these nephrogenic rests are divided into intralobar rests (i.e., within the lobe of the kidney) or perilobar rests (i.e., at the periphery of the kidney). Perilobar nephrogenic rests are associated with 11p15 (*WT2*): Beckwith-Wiedemann syndrome and synchronous bilateral Wilms' tumor. Intralobar nephrogenic rests are associated with 11p13 (*WT1*): aniridia, Denys-Drash syndrome,

and metachronous Wilms' tumor .

| **Overgrowth** | **Non-overgrowth** |
|---|---|
| Beckwith-Wiedemann | Denys-Drash |
| Hemihypertrophy | WAGR |
| Perlman | Aniridia |
| Sotos | |

# PATHOLOGY OF WILMS' TUMOR

## NWTS-5 INSTITUTIONAL PATHOLOGY CHECKLIST

COMPLETE OR PARTIAL NEPHRECTOMY SPECIMENS ONLY

Checklist to be completed by pathologist of record.

Patient name: _____ Patient ID#: ☐☐☐☐☐☐

Institution: _____

Surgical pathology specimen number(s): _____

1. Specimen weight: (gm.): _____
   Weight of removed kidney and associated tumor in grams:

2. Source of specimen: ( )Pre-treatment ( )Post-treatment

3. Type of specimen: (Complete **two forms** whenever tissue is available from both kidneys)
   ( )Unilateral, complete nephrectomy ( )Left nephrectomy or partial nephrectomy (bilateral)
   ( )Right nephrectomy or partial nephrectomy (bilateral)

4. Largest tumor diameter (cm.): ___ ___ ___
   For multicentric tumors, indicate the diameter of the largest single tumor.

5. Specimen received intact and unopened from O.R.? ( ) No ( )Yes ( )Uncertain

6. Renal capsule grossly intact? Before opening specimen? ( )No ( )Yes ( )Uncertain

   Comments: _____

7. Surface inked? Before opening specimen? ( ) No ( )Yes ( )Uncertain
   After opening specimen? ( ) No ( )Yes ( )Uncertain

8. Tumor multicentric? ( ) No ( )Yes ( )Uncertain

   Comments: _____

9. Margin involved by tumor? ( ) No ( ) Yes ( ) Uncertain
   Comments: _____

10. Regional nodes (hilar, periaortic or other abdominal sites):
    ( ) Negative for tumor ( ) Positive for tumor ( ) Uncertain ( ) None examined

    Comments: _____

***Please Complete Sections 11, 12, And 13 Based Upon Your Diagnosis Before NWTSG Review***

11. Your diagnosis (before NWTSG review): _____
    ( )Mesoblastic nephroma ( )FH ( )Focal Anaplasia ( )Diffuse Anaplasia ( )Anaplasia,NOS
    ( )Clear Cell ( )Rhabdoid ( )Other Sarcoma ( )Nephroblastomatosis only
    ( )Other, specify: _____
    ( )Uncertain or Unknown, specify: _____

12. For unilateral case: was contralateral biopsy obtained? ( ) No ( ) Yes
    If yes, result: ( ) Normal tissue ( ) Nephrogenic rest ( ) Uncertain



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Patient ID#: ☐☐☐☐☐☐

13. Local stage based on pathological exam. For bilateral cases, indicate local stage of side for which this form is completed. **Specify local stage even for Stage IV cases**.

( ) I
( ) II Reason(s):_____
( ) III Reason(s):_____

Comments: _____

Check all staging criteria below that apply:

**STAGE II**

( )Tumor penetrates renal capsule into perirenal fat
( )Tumor capsule biopsied prior to nephrectomy without diffuse peritoneal soilage (including a needle biopsy)
( )Tumor involves blood or lymphatic vessels within the renal sinus
( )Tumor in main renal vein apparently removed without cutting across tumor
( )Tumor infiltrated adjacent organs or vena cava, but is completely resected
( )No tumor in nodes
( )Margins of specimen free of tumor

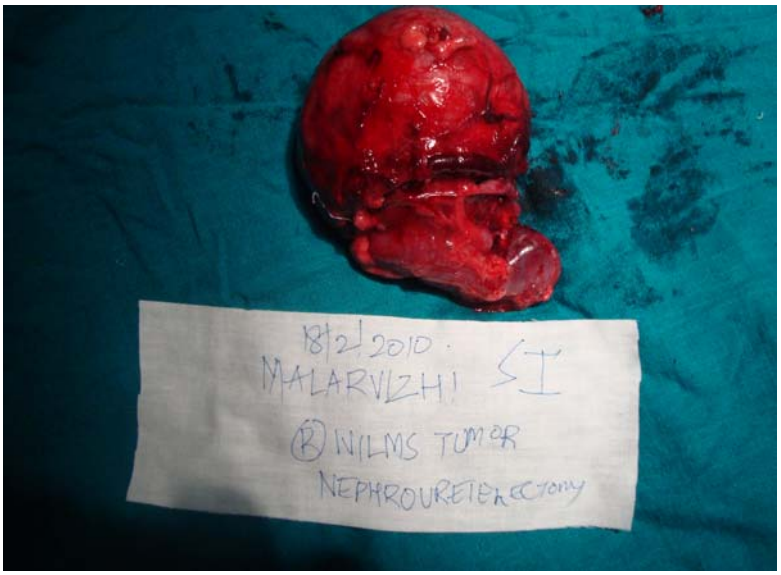
**STAGE III**

( )Surgical margins involved by tumor, with known or presumed residual disease
( )Tumor rupture or biopsy with peritoneal soilage
( )Tumor in lymph nodes
( )Tumor thrombi transected or removed piecemeal by surgeon
( )Peritoneal implants present
( )Tumor incompletely resected

Form completed by: _____ (Signature) Date: _____/_____/_____ (mm / dd / yy)

Printed name: _____

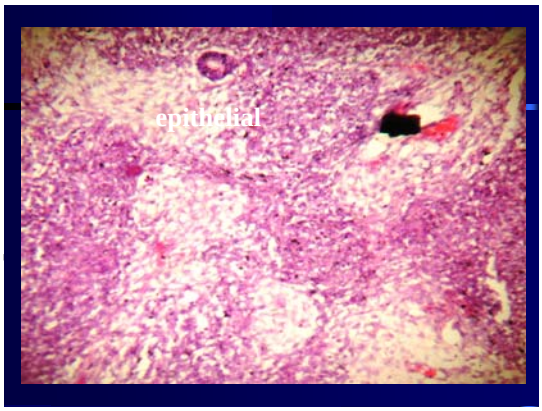
## PATHOLOGY OF WILMS TUMOUR



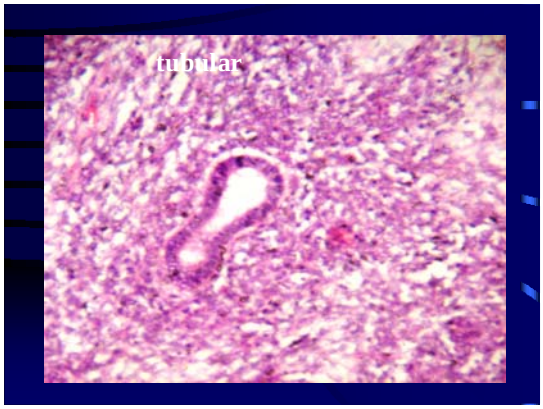

On gross specimen, Wilms' tumor has a varied appearance of smooth to cystic and variegated on cut section. There is no distinct capsule, but the surrounding mesenchyme condenses to form a pseudocapsule. Occasionally, hemorrhage and necrosis are noted on gross specimen examination. The whole specimen is handled very carefully, and gross specimen examination, microscopic study, and further biologic studies are performed according to the protocols by SIOP 2002 in Europe and the American College of Pathologists in the United States. For macroscopic examination, the tumor should be send to the pathology department intact without formalin preservative. The examination includes specimen weight, tumor location, capsule invasion, renal vein and sinus invasion, ureter, and cut surface of the kidney if a heminephrectomy was performed.

**We have had 154 favourable histology and two unfavourable in our study**

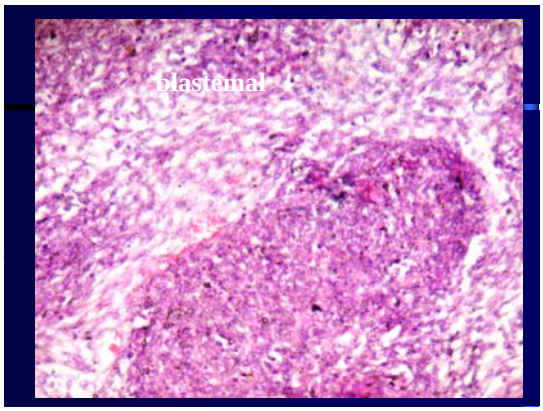
**EPITHELIAL**




**TUBULAR**




**BLASTEMAL**



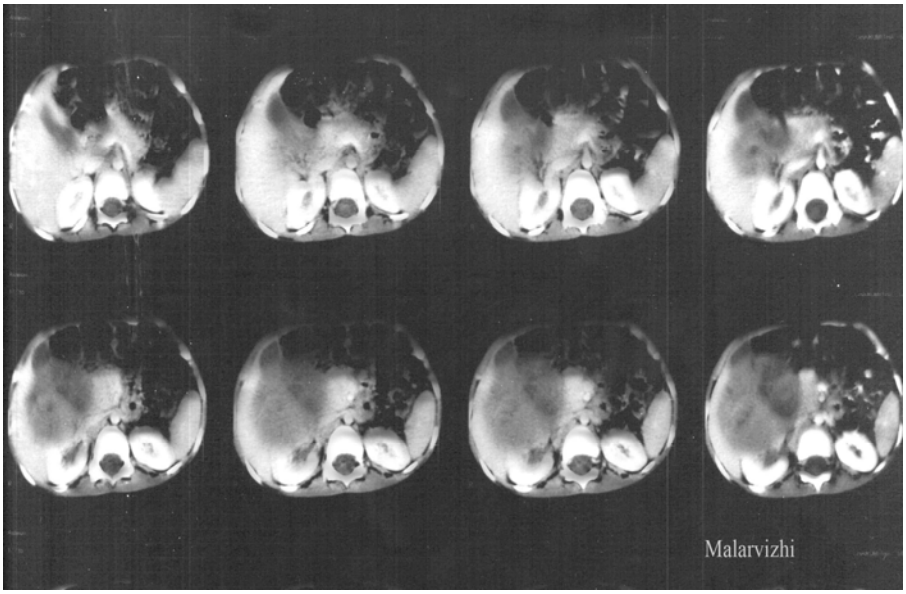
## MANAGEMENT OF WILMS' TUMOR


**IVU-shows calyceal distortion in right kidney in wilms tumour**

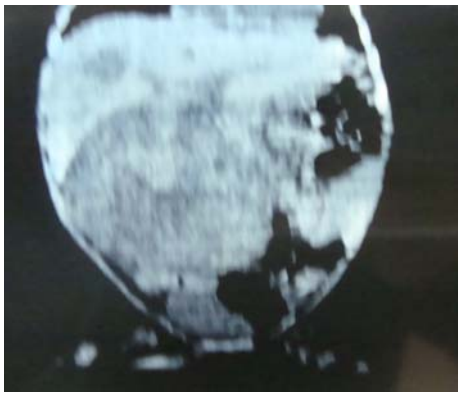
3year male child presented with lung secondaries and right kidney involved with wilms tumour.

A computed tomography scan of a 3.5-year-old girl who presented with a large abdominal mass demonstrating the characteristic findings of

a Wilms' tumor. The tumor mass can be seen protruding from the right kidney with a margin of renal parenchyma along the periphery.







Rhabdoid tumour with liver metastases

Management consists of surgery for removal of the primary tumor with the kidney (radical nephrectomy), with chemotherapy and radiotherapy in some cases. The management is guided by the NWTS

protocol in ICH-CHENNAI and in the United States and the SIOP protocol in Europe according to the stage of the tumor.

**Surgery**

Radical nephrectomy for removal of the primary tumor with the kidney is the mainstay of treatment. This procedure allows the removal of the primary tumor and accurate staging of the tumor. The usual approach is transperitoneal through a transverse abdominal incision, which gives good access to the tumor and vasculature.

**The principles of surgery are as follows:**

1. Palpation of liver, abdomen, and para-aortic region for regional spread of disease

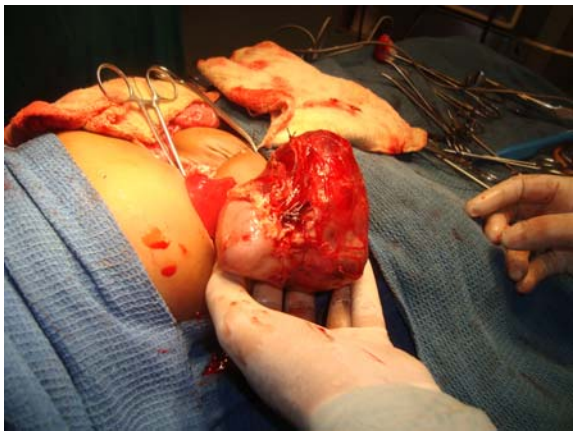
2. Removal of intact specimen in total

3. Avoidance of local spillage because these children have a sixfold increase in local abdominal relapse[36]

4. Nodal sampling rather than clearance because there is no added advantage in the long-term survival[37]

5. Proper identification and avoidance of injury to contralateral renal vessels, aorta, and iliac and superior mesenteric arteries[38]

6. Palpation of the renal vein and inferior vena cava before ligation to rule out thrombus.



# PRINCIPLES OF RADICAL NEPHRO URETERECTOMY

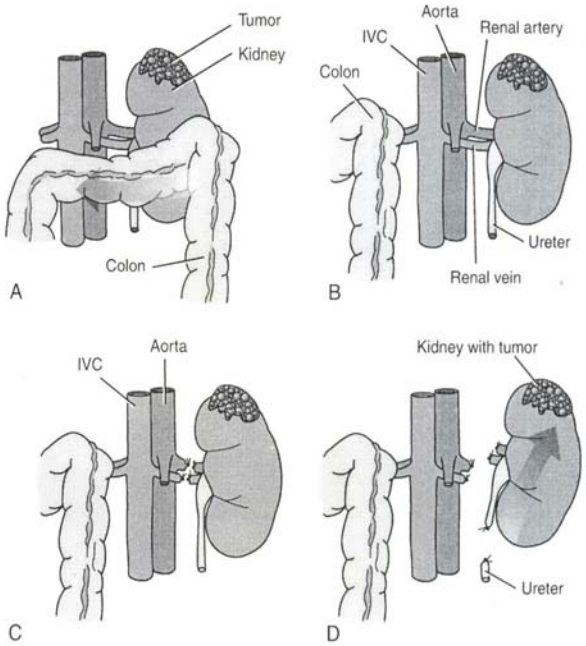



**Figure 50-8** Surgical steps showing tumor nephrectomy on left side. **A,** Reflection of colon from kidney. **B,** Identification of major vessels and control at renal hilum. **C,** Ligation of renal vessels. **D,** Removal of tumor with kidney, and ligation of ureter. IVC, inferior vena cava.

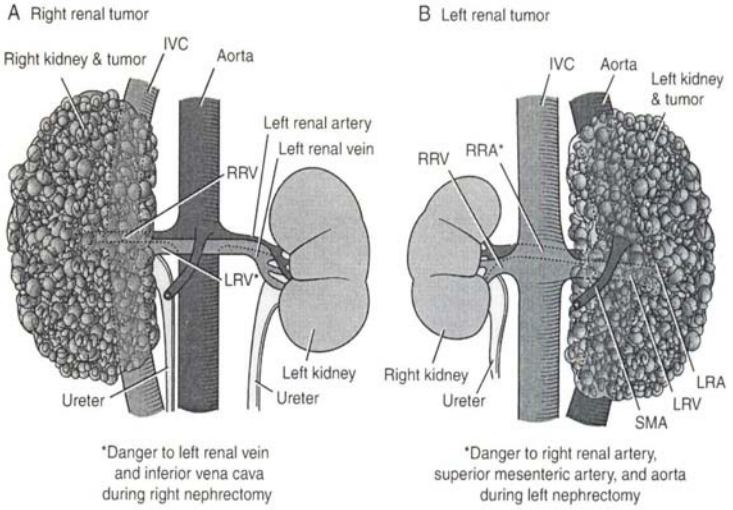



**Figure 50-9** Risk of vascular injury during right **(A)** and left **(B)** renal tumor nephrectomy. IVC, inferior vena cava; LRA, left renal artery; LRV, left renal vein; RRA, right renal artery; RRV, right renal vein; SMA, superior mesenteric artery.

Primary Nephrectomy (NWTS) and Neoadjuvant Chemotherapy Followed by Nephrectomy (SIOP)GEARHART-Pediatric urology.

According to the NWTS protocol, following confirmation on imaging, the nephrectomy is performed enabling the collection of tissue for histologic diagnosis, primary removal of the tumor, and exact staging. This surgery is often challenging, however, in view of the tumor size and the need to adhere to strict principles of surgery, especially regarding intraoperative tumor spilling.

According to the SIOP protocol, after imaging of the tumor, a fine-needle biopsy is performed to obtain a histologic diagnosis. Under the same anesthesia, vascular access (Hickman line) is obtained for administering the chemotherapy. After 4 weeks of chemotherapy, the response is assessed by repeat ultrasound scan, and nephrectomy is performed on week 5 or 6 (SIOP-9 2001). The potential advantage is that the tumor is small, and the risk of intraoperative spillage is reduced.

According to the UKW3 study group, this approach has also resulted in a shift toward a more advantageous stage distribution and reduction in therapy and late effects of doxorubicin and radiotherapy.[44] The disadvantage is that studying tumor specimen histology after chemotherapy may not be the same as the NWTS protocol, especially nodal histology for defining postoperative chemotherapy.

**Indications of Preoperative Chemotherapy for Wilms' Tumor (NWTS)**

(Gearhart-50-677) primary nephrectomy is performed according to the NWTS protocol, but there is a role for preoperative chemotherapy in patients with the following conditions:

1. Inoperable tumor at the primary nephrectomy because removal of surrounding organs can result in an increased risk of surgical complications

2. Inferior vena cava thrombus, especially the suprahepatic level, reducing the need for cavotomy and cardiopulmonary bypass as per NWTS and UKW3 trial

3. Bilateral tumor at the diagnosis or incidental finding, at the primary nephrectomy.

***Surgical Complications***

- NWTS has reported a surgical complication rate of 11% for primary nephrectomy.

- Common complications were hemorrhage and small intestinal obstruction. Other complications are intussusception and damage to adjacent organs.

- The cause of intestinal obstruction in the late postoperative period is mostly adhesions. Risk factors identified were higher local tumor stage, intravascular extension, and

- en bloc resection of other visceral organs

# MANAGEMENT OF BILATERAL WILMS TUMOUR

Our management in bilateral wilms tumor includes incisional biopsy and confirmation. After confirmation , chemotherapy was given for 6 cycles with Vincristine , pulse-intensive Actinomycin D. After 6 cycles of chemotherapy, surgery was undertaken .Out of 9 patients, 7 patients underwent unilateral radical nephrectomy and 2 partial nephrectomy Further 6 cycles of chemotherapy was given For the residual lesion in opposite kidney, radiotherapy of 10.8Gy was given in 16 sittings .Disease free survival years are less than 2 years.

# MATERIALS AND METHODS

## Patients and Methods

It is a combined prospective and retrospective study which pediatric urology OPD at the Institute of Child Health and Hospital for Children, Madras Medical College, Chennai. The study included patients with wilms, who attended the pediatric surgery during the ten years period, from Jan 1, 2000 to Dec 31, 2009.

### Selection Criteria

All patients with renal mass, hematuria, fever, abdominal pain

### Inclusion Criteria

All patients with wilms tumour were proven radiologically , sonographically and HPE wise

### Exclusion Criteria

- stromal tumours as clear cell sarcoma .
- congenital mesonephric hamartoma
- rhabdoid tumour.
- angiomyolipoma of the kidney

The patients were subjected to detailed clinical examination and relevant investigations were performed, namely, ultrasound examination, IVU, +/- CECT.The treatment modalities were studied and patients were followed up to screen for residual lesion size, recurrences and liver and lung secondaries.

# PROFORMA

Personal Details -

Name Age Sex

IP No.

Address

Date

## Clinical Symptoms

□ Asymptomatic □ abdominal mass

□ Haematuria □ anemia

□ hypertension □ dysuria

□ aniridia

## Laterality

□ U/L (Rt/Lt)

□ B/L-

## Investigations

**Renal Parameters**

BU/SC/SE –

**Contra-lateral Renal Status (in U/L cases and other problems) –**

U/S KUBU Report –mass cystic or solid, presence of thrombus in IVC or renal vein

**IVU – calyceal distortion, duplex system is well seen in ivu than CECT**

CECT –confimative of mass lesion -

## Treatment

□ Primary radical nephro-ureterectomy followed by chemotherapy

□ radiotherapy for stageII, III, IV, and V

**HPE Report**

Favorable triphasic histology.

Unfavorable anaplastic

Focal versus diffuse anaplasia

**post-op follow up monthly f/u**

U/S abdomen : look for residual and recurrent lesion and secondaries in liver

**Chest xray : look for secondaries in lungs**

**Follow-up protocol**

- monthly f/u  for 6months
- 3monthly f/u  for 6visits.
- 6monthly f/u for 2years
- yearly f/u  till life

# MASTER CHART

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| Jerusha | 2/F | 14.01.03 | 5 | B/L | FH | Triphasic | PRE-OP CHEMO18WKS | Ct&Rt | L.NU.RK-N | 18.3.03 |
| Kani | 3/m | 4.11.03 | 5 | B/L | FH | Triphasic | Partial nephrectomyB/L | CT&RT | residual+ | 10.08.04 |
| Ramesh | | 12.11.03 | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Sajad | | | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Vimalraj | 1/M | | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Monisha | 2y6m | 2007 | 5 | B/L | FH | Triphasic | l.nephroureterectomy &rt.debulking-610679 | CT&RT | | Supraclavicular node+ |
| Anitha | 1y6m | 3.04.07 | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Veeraraj | 7/m | 15.11.99 | 5 | B/L | FH | Triphasic | l.nephroureterectomy &rt.debulking-610679 | CT&RT | anuria | 15.4.03 |
| Benedictroselin- | 4/m | 2008 | 5 | B/L | FH | Triphasic | chemo-l.nephrouret erectomy-RT | CT&RT | | |
| jayasoorya | 3y6m | 13.6.06 | 2 | R | focal anaplasia | Triphasic | r.nephroureterectomy | CT&RT | | 2009-DEC |
| Sathish | 3/m | | 2 | R | residual r.wt | Triphasic | r.nephroureterectomy | CT&RT | | 2008-AUG |
| Kanya | 1y | | 1 | R | FH | Triphasic | metachronous WT | CT&RT | lung sec. | |
| kamatchi | 8/f | | 4 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | liver sec | |
| Dinesh | 3/m | 20.06.07 | 2 | R | FH | Triphasic | metachronous WT | CT&RT | ascites | |
| Saranya | 11/f | 2000TMC | 4 | L | FH | Triphasic | liver sec | CT&RT | ascites | 2006-f/u |
| Ramya | <1/f | | 1 | R | FH | Triphasic | metachronous WT | CT&RT | r.lung sec | |
| ramachandran | 8/m | Feb-97 | 3 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | | 2006-well |
| Karthik | 6/mm | jan-98-madurai | 3 | R | FH | Triphasic | cystic wilms | CT&RT | tumour spill | 2000-AMA |
| Aravind | 2y6m | | 3 | L | FH | Triphasic | L.nephroureterectomy | CT&RT | | |
| anbarasan | 1/m | | 1 | L | FH | Triphasic | l.nephroureterectomy | | | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| beula | 9/f | 17.10.98 | 1 | L | FH | Triphasic | l.nephroureterectomy | CT&RT | | 2003 |
| babu | 10/m | | 3 | Horseshoe kidney-03 | FH | Triphasic | partial nephrectommmmy | CT&RT | | 2003 |
| bavani | 5/f | | 3 | L | FH | Triphasic | l.nephroureterectomy | | | |
| Elizabeth | 4/f | 1997 | 3 | L | FH | Triphasic | L.nephroureterectomy | | | |
| hakeem | | | 1 | L | FH | Triphasic | L.nephroureterectomy | | | 2005 |
| hemanathan | | | 1 | L | FH | Triphasic | L.neprhroureterectomy | | | 2005 |
| Habibunisha | 8/f | | 1 | L | FH | Triphasic | l.nephroureterectomy | | | |
| janathnisha 7month | 7months | | 2 | R | FH | Triphasic | r.nephroureterectomy | | | 2006 |
| jagadesh | 4/m | | 3 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | | |
| Jayaraman | 2/m | | 1 | R | FH | triphasic | r.nephroureterectomy | hemihypertrophy | | 2009 |
| jagan | | | 1 | L | | triphasic | r.nephroureterectomy | | | 2003 |
| karthik | 4/m | | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| kousalya | 1/f | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| kalaiselvi | 1/f | | 2 | L | FH | triphasic | r.nephroureterectomy | | | |
| kavinraj | 1/f | 12.01.02 | 1 | R | FH | triphasic | r.nephroureterectomy | | | f/u3.08.08 |
| lavanya | 6/f | Nov-01 | 2 | L | FH | triphasic | l.nephroureterectomy | 366277 | | f/u-2003 |
| mohaprabu | 3months | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| manojkum | 5/m | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| manirathnam | 4/m | | 1 | L | FH | triphasic | l.nephroureterectomy | | | |
| malarvizhi | 5months | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| manimegalai 1/f | | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| nandagopal | 1/m | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| nithya | 9/f | 5.1.99 | 2 | L | FH | triphasic | l.nephroureterectomy | 264470 | | |
| Priya | | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| priyadarshini | | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| prescila | 3y6 | | 3 | L | FH | triphasic | l.nephrouretectomy | terectomy | | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| preethy | 3y6 | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| sudhakar | 4y6 | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| satyanarayan | 3/m | | 2 | R | FH | triphasic | r.nephroureterectomy | | | 2010-may |
| Satish | 3/m | | 3 | R | FH | triphasic | r.nephroureterectomy | | | |
| suresh | 2/m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | May-10 |
| vijay | 3/m | | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | |
| vimalraj | 7months | May-01 | 2 | R | FH | triphasic | r.nephroureterectomy | half dosechemo | | |
| vignesh | 7/m | | | L | FH | triphasic | l.nephroureterectomy | | | |
| vino | 5/m | | 3 | R | FH | triphasic | r.nephroureterectomy | | | |
| vanitha | 5/f | Dec-00 | 4 | R | FH | triphasic | r.nephroureterectomy | lung sec-mar.01 | | 2003-well |
| yuvarani | 3/f | | | R | FH | triphasic | r.nephroureterectomy | | | |
| naveenkumar | 1y6m | | | L | FH | triphasic | l.nephroureterectomy | | | |
| praveena | 4/f | | 3 | R | FH | tubullar | r.nephroureterectomy | | | |
| chelladurai 1/m | | | 3 | L | FH | triphasic | L.nephrourterectomy | | | |
| pushpakar | 3/m | May-02 | 3 | L | FH | triphasic | L.nephroureterectomy | CT&RT | ?cxr-sec | 30.09.08 |
| aravind | 2y6m | f/u-30.03.04 | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| ajitkumar | 4/m | 21.2.04 | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| archana | 5/f | 09.09.04 | 3 | L | FH | triphasic | l.nephrourreterectomy | | | Dec-09 |
| adinarayanan | 7/m | Mar-04 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Feb-10 |
| dilliprakash | 1y3m | 19.10.05 | 3 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Jan-09 |
| franciskennady | | Jan-03 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Dec-09 |
| gowthami | 5/f | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Apr-10 |
| Hemadevi | 1y9m | 23.03.04 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | May-10 |
| ilakya | 2/f | 30.06.05 | 1 | L | FH | triphasic | l.nephroureterectomy | defaulted | | |
| Jayashree | 1y6m | 28.10.03 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Nov-09 |
| kavya | 1y3m | 2003 | 3 | R | FH | epithelial | r.nephroureterectomy | CT&RT | | Aug-09 |
| Kumaran | 3y6m | 19.10.04 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| karthik | 3/m | jan.05 | 1 | R | FH | triphasic | r.nephroureterectomy | CT&RT | recurrence | Sep-09 |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| karthik | 4/m | jan.05 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Oct-09 |
| lokesh | 1O/M | 21.06.05 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| sandya | 4/f | 2009 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jun-09 |
| Manikandan | 3/m | 29.3.03 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Nov-09 |
| Manikandan | 3/m | 19.1.04 | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| Meenakshi | 6/F | 27.03.04 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Jan-10 |
| nithya | 9/f | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| dathathreyan | 1y6m | 2008 | 3 | R | FH | triphasic | r.nephroureterectomy | | | Dec-09 |
| Pooja | 1Y1M | 2008 | 1 | L | FH | blastemal | l.nephroureterectomy | | | Feb-10 |
| sindhu | 5/m | 2008 | 1 | R | FH | epithelial | r.nephroureterectomy | | | Apr-10 |
| reddiyama | 5/f | 2008 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Mar-10 |
| chandru | 1y6m | 2008 | 1 | R | FH | triphasic | r.nephroureterectomy | | | May-10 |
| dhanusha | 4/f | 2008 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Dec-09 |
| wahithabanu | 4/f | 2008 | 3 | L | FH | triphasic | l.nephroureterectomy | | | Nov-09 |
| abimanu | 4/m | 13.06.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | Jun-09 |
| Anitha | 1y6m | 3.04.07 | 5 | b/l | FH | triphasic | | CT&RT | | lostf/u |
| barath | 10months | 4.12.07 | 1 | L | FH | triphasic | l.nephroureterectomy | | | Aug-09 |
| Darshini | 1y6m | 10.07.07 | 1 | R | FH | triphasic | r.nephroureterectomy | | | May-10 |
| dinesh | 10months | 30.04.07 | 1 | L | FH | triphasic | l.nephroureterectomy | | | Apr-09 |
| dathatreyan | 1y6m | 24.01.08 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| gouthami | 6/f | Jun-04 | 3 | R | FH | triphasic | r.nephroureterectomy | thrombus not extracted | | f/u-24.5.08 |
| ganesan | 1y6m | 26.06.06 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Sep-09 |
| gayathri | 2y6m | 11.07.06 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Oct-09 |
| harshavardan | 4/m | 10.06.06 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jan-10 |
| kamesh | 3y6m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jul-09 |
| kalaiselvan | 6/m | | 3 | L | FH | triphasic | | CT&RT | | lostf/u |
| monisha | 4y6m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| niranjan | | Mar-06 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Feb-09 |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| parthiban | 2y9m | 19.01.06 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Aug-09 |
| purosathaman | 5/m | 18.07.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | Apr-10 |
| praveen | 7/m | | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Mar-10 |
| pooja | 1/f | 19.09.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| pooja | 11months | 18.01.08 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| Ravikumar | 4/m | 26.10.06 | 1 | L | FH | triphasic | L.nephroureterectomy | | | 16.9.08 |
| tamilarasan | 1y6m | 30.12.06 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| tangamani | 3/m | 13.06.07 | 2 | R | FH | triphasic | r.nephroureterectomy | local recurrence | | lostf/u |
| anish | 11months | 2008 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Apr-10 |
| pavithra | 2/f | 2008 | 1 | L | FH | triphasic | L.nephroureterectomy | cysticwilm | | May-10 |
| mohanraj | 3/m | 2008 | 2 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Sep-09 |
| sudharsana | 7/m | 1.7.03 | 3horse shoe kidney | | FH | triphasic | L.nephroureterectomy | tumour spill | | f/u-23/5/06 |
| pushpakar | 4y6m | 2003 | 2 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Jul-09 |
| pavithra | 2/f | 2003 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jun-09 |
| parthiban | 1y6m | 2003 | 1 | L | FH | triphasic | l.nephroureterectomy | | | |
| syedsulthana | 5mon | 2003 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| sheela | 4/f | 2003 | 2 | L | FH | triphasic | L.nephroureterectomy | ct&rt | | Feb-20 |
| sharmila | 3y6m | 2004 | 1 | R | FH | triphasic | r.nephroureterectomy | 522799 | 2008-well | |
| sundarrajan | 8/m | 2005 | 2 | R | UFH | triphasic | r.nephroureterectomy | | | lost f/u |
| surya | 5/m | 2005 | duplex-2 | R | FH | triphasic | r.heminephrectomy | 446864 | recurrence | defaultedRT |
| sakthivel | 10months | 2006 | 4 | L | FH | triphasic | L.nephroureterectomy | | | Oct-09 |
| Thirumalai | 4/m | 2006 | 2 | L | FH | triphasic | L.nephroureterectomy | | | Nov-09 |
| thirunavukarasu | 9/m | 2006 | 2 | L | FH | triphasic | L.nephroureterectomy | | | Jun-09 |
| thilani | 5/f | 2005 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Sep-08 |
| thirumurugan | 5/m | 1.2.2006 | 3 | R | FH | triphasic | r.nephroureterectomy | 524468 | f/u9.9.08 | |
| vasanthkumar | 7/m | Mar-06 | | L | FH | triphasic | L.nephroureterectomy | 506170 | | Sep-09 |
| yuvaraj | 1y6m | Sep-06 | | L | FH | triphasic | L.nephroureterectomy | | | Jan-10 |
| yasmin | 4/f | Apr-06 | | L | FH | triphasic | L.nephroureterectomy | | | Apr-09 |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| hemadevi | 1y9m | 3.4.04 | 2 | R | FH | triphasic | r.nephroureterectomy | | | Feb-10 |
| lavanya | 8/f | 23.02.05 | 3 | R | FH | triphasic | r.nephroureterectomy | lung RT-givenf/u-15.7.08 | | |
| kaviarasan | | 1997 | | L | FH | triphasic | L.nephroureterectomy | | 2008-well | |
| sangeetha | | | 3 | L | FH | triphasic | L.nephroureterectomy | 8yr-survival | | |
| soubarnika | | 97 | | R | FH | triphasic | r.nephroureterectomy | 11yr-survival | | |
| sathishkumar | | | | R | FH | triphasic | r.nephroureterectomy | 15yr-survival | | |
| Binnsajan | 6/m | 568257 | 2 | L | FH | triphasic | l.nephroureterectomy | recurrence | lung sec | |
| kamurunisha- | 12/f | | 2 | L | FH | triphasic | l.nephroureterectomy | hemihypertrophy | | May-10 |
| muthumadhavan | 6/m | | duplex-3 | | FH | triphasic | l.nephroureterectomy | | | |
| thanusha | 4/f | | 3 | R | FH | triphasic | atrial thrombus-partial thrombectomy | | | |
| kumaran | 3y6m | 473179 | 3 | R | FH | triphasic | l.nephroureterectomy | thrombectomy | | f/u2009-feb |
| chaitanya | 3/f | | 3 | R | FH | triphasic | tumor dissemination | | neoadjuvani | |
| rohith | 2/m | | 3 | L | FH | triphasic | l.nephroureterectomy | biopsy | death | |
| malarvizhi | 5/f | 674021 | 1 | R | FH | triphasic | r.nephroureterectomy | hpe195/10 | | |
| deepika | | | 3 | L | FH | epithelial | l.nephroureterectomy | | neoadjuvant | |
| sudharsan | 1y6m | 672054 | | R | FH | triphasic | r.nephroureterectomy | | | |
| deepak | 2/m | 674525 | | l | FH | triphasic | l.nephroureterectomy | | | |
| sabarivasan- | 2/m | 673 | 2 | l | FH | triphasic | l.nephroureterectomy | | WAGR-syndrome | |
| monisha | 2/F | 674101 | 4 | R | FH | Rhabdomyomatous | r.nephroureterectomy | | neoadjuvant | |
| soundarya | 6/f | 673833 | 3 | R | FH | Rhabdomyomatous | r.nephroureterectomy | ascites | neoadjuvant | |
| aswathdevi | 1y6m | 676082 | 3 | R | FH | Rabdomyomatous | l.nephroureterectomy | | neoadjuvant | |
| jeganbabu | 4M | 664680 | | L | FH | | r.nephroureterectomy | | | |
| harish | 3y6m | 658953 | 4 | R | FH | triphasic | r.nephroureterectomy | lung sec | death | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| nivedha | 3/F | | 2 | L | FH | triphasic | l.nephroureterectomy | | | Apr-10 |
| alifa | 9months | | 4 | L | UFH | rhabdoid | l.nephroureterectomy | liver sec | death | |
| senbagavel | 3/m | | 3 | R | FH | triphasic | | | neoadjuvant | |
| Jerusha | 2/f | 14.01.03 | 5 | B/L | FH | Triphasic | pre-op chemo18wks | CT&RT | l.nu.rk-N | 18.3.03 |
| Kani | 3/m | 4.11.03 | 5 | B/L | FH | Triphasic | Partial nephrectomyB/L | CT&RT | residual+ | 10.08.04 |
| Ramesh | | 12.11.03 | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Sajad | | | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Vimalraj | 1/M | | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Monisha | 2y6m | 2007 | 5 | B/L | FH | Triphasic | l.nephroureterectomy &rt.debulking-610679 | CT&RT | | Supraclavicular node+ |
| Anitha | 1y6m | 3.04.07 | 5 | B/L | FH | Triphasic | neo-adjuvant | CT&RT | | |
| Veeraraj | 7/m | 15.11.99 | 5 | B/L | FH | Triphasic | l.nephroureterectomy &rt.debulking-610679 | CT&RT | anuria | 15.4.03 |
| Benedictroselin- | 4/m | 2008 | 5 | B/L | FH | Triphasic | chemo-l.nephrouret erectomy-RT | CT&RT | | |
| jayasoorya | 3y6m | 13.6.06 | 2 | R | focal anaplasia | Triphasic | r.nephroureterectomy | CT&RT | | 2009-DEC |
| Sathish | 3/m | | 2 | R | residual r.wt | Triphasic | r.nephroureterectomy | CT&RT | | 2008-AUG |
| Kanya | 1y | | 1 | R | FH | Triphasic | metachronous WT | CT&RT | lung sec. | |
| kamatchi | 8/f | | 4 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | liver sec | |
| Dinesh | 3/m | 20.06.07 | 2 | R | FH | Triphasic | metachronous WT | CT&RT | ascites | |
| Saranya | 11/f | 2000TMC | 4 | L | FH | Triphasic | liver sec | CT&RT | ascites | 2006-f/u |
| Ramya | <1/f | | 1 | R | FH | Triphasic | metachronous WT | CT&RT | r.lung sec | |
| ramachandran | 8/m | Feb-97 | 3 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | | 2006-well |
| Karthik | 6/mm | jan-98-madurai | 3 | R | FH | Triphasic | cystic wilms | CT&RT | tumour spill | 2000-AMA |
| Aravind | 2y6m | | 3 | L | FH | Triphasic | L.nephroureterectomy | CT&RT | | |
| anbarasan | 1/m | | 1 | L | FH | Triphasic | l.nephroureterectomy | | | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| NAME | Age & sex | F/U | STAGE | | HPE | HPE | Surgery | Chemo & RT | | last f/u |
| beula | 9/f | 17.10.98 | 1 | L | FH | Triphasic | l.nephroureterectomy | CT&RT | | 2003 |
| babu | 10/m | | 3 | Horseshoe kidney-03 | FH | Triphasic | partial nephrectommmmy | CT&RT | | 2003 |
| bavani | 5/f | | 3 | L | FH | Triphasic | l.nephroureterectomy | | | |
| Elizabeth | 4/f | 1997 | 3 | L | FH | Triphasic | L.nephroureterectomy | | | |
| hakeem | | | 1 | L | FH | Triphasic | L.nephroureterectomy | | | 2005 |
| hemanathan | | | 1 | L | FH | Triphasic | L.neprhroureterectomy | | | 2005 |
| Habibunisha | 8/f | | 1 | L | FH | Triphasic | l.nephroureterectomy | | | |
| janathnisha 7month | 7months | | 2 | R | FH | Triphasic | r.nephroureterectomy | | | 2006 |
| jagadesh | 4/m | | 3 | R | FH | Triphasic | r.nephroureterectomy | CT&RT | | |
| Jayaraman | 2/m | | 1 | R | FH | triphasic | r.nephroureterectomy | hemihypertrophy | | 2009 |
| jagan | | | 1 | L | | triphasic | r.nephroureterectomy | | | 2003 |
| karthik | 4/m | | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| kousalya | 1/f | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| kalaiselvi | 1/f | | 2 | L | FH | triphasic | r.nephroureterectomy | | | |
| kavinraj | 1/f | 12.01.02 | 1 | R | FH | triphasic | r.nephroureterectomy | | | f/u3.08.08 |
| lavanya | 6/f | Nov-01 | 2 | L | FH | triphasic | l.nephroureterectomy | 366277 | | f/u-2003 |
| mohaprabu | 3months | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| manojkum | 5/m | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| manirathnam | 4/m | | 1 | L | FH | triphasic | l.nephroureterectomy | | | |
| malarvizhi | 5months | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| manimegalai 1/f | | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| nandagopal | 1/m | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| nithya | 9/f | 5.1.99 | 2 | L | FH | triphasic | l.nephroureterectomy | 264470 | | |
| Priya | | | 2 | R | FH | triphasic | r.nephroureterectomy | | | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| priyadarshini | | | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| prescila | 3y6 | | 3 | L | FH | triphasic | l.nephrouretectomy | terectomy | | |
| preethy | 3y6 | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| sudhakar | 4y6 | | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| satyanarayan | 3/m | | 2 | R | FH | triphasic | r.nephroureterectomy | | | 2010-may |
| Satish | 3/m | | 3 | R | FH | triphasic | r.nephroureterectomy | | | |
| suresh | 2/m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | May-10 |
| vijay | 3/m | | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | |
| vimalraj | 7months | May-01 | 2 | R | FH | triphasic | r.nephroureterectomy | half dosechemo | | |
| vignesh | 7/m | | | L | FH | triphasic | l.nephroureterectomy | | | |
| vino | 5/m | | 3 | R | FH | triphasic | r.nephroureterectomy | | | |
| vanitha | 5/f | Dec-00 | 4 | R | FH | triphasic | r.nephroureterectomy | lung sec-mar.01 | | 2003-well |
| yuvarani | 3/f | | | R | FH | triphasic | r.nephroureterectomy | | | |
| naveenkumar | 1y6m | | | L | FH | triphasic | l.nephroureterectomy | | | |
| praveena | 4/f | | 3 | R | FH | tubullar | r.nephroureterectomy | | | |
| chelladurai 1/m | | | 3 | L | FH | triphasic | L.nephrourterectomy | | | |
| pushpakar | 3/m | May-02 | 3 | L | FH | triphasic | L.nephroureterectomy | CT&RT | ?cxr-sec | 30.09.08 |
| aravind | 2y6m | f/u-30.03.04 | 3 | L | FH | triphasic | l.nephroureterectomy | | | |
| ajitkumar | 4/m | 21.2.04 | 2 | R | FH | triphasic | r.nephroureterectomy | | | |
| archana | 5/f | 09.09.04 | 3 | L | FH | triphasic | l.nephrourreterectomy | | | Dec-09 |
| adinarayanan | 7/m | Mar-04 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Feb-10 |
| dilliprakash | 1y3m | 19.10.05 | 3 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Jan-09 |
| franciskennady | | Jan-03 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Dec-09 |
| gowthami | 5/f | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Apr-10 |
| Hemadevi | 1y9m | 23.03.04 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | May-10 |
| ilakya | 2/f | 30.06.05 | 1 | L | FH | triphasic | l.nephroureterectomy | defaulted | | |
| Jayashree | 1y6m | 28.10.03 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Nov-09 |
| kavya | 1y3m | 2003 | 3 | R | FH | epithelial | r.nephroureterectomy | CT&RT | | Aug-09 |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| Kumaran | 3y6m | 19.10.04 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| karthik | 3/m | jan.05 | 1 | R | FH | triphasic | r.nephroureterectomy | CT&RT | recurrence | Sep-09 |
| karthik | 4/m | jan.05 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Oct-09 |
| lokesh | 1O/M | 21.06.05 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| sandya | 4/f | 2009 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jun-09 |
| Manikandan | 3/m | 29.3.03 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Nov-09 |
| Manikandan | 3/m | 19.1.04 | 2 | L | FH | triphasic | l.nephroureterectomy | | | |
| Meenakshi | 6/F | 27.03.04 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Jan-10 |
| nithya | 9/f | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| dathathreyan | 1y6m | 2008 | 3 | R | FH | triphasic | r.nephroureterectomy | | | Dec-09 |
| Pooja | 1Y1M | 2008 | 1 | L | FH | blastemal | l.nephroureterectomy | | | Feb-10 |
| sindhu | 5/m | 2008 | 1 | R | FH | epithelial | r.nephroureterectomy | | | Apr-10 |
| reddiyama | 5/f | 2008 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Mar-10 |
| chandru | 1y6m | 2008 | 1 | R | FH | triphasic | r.nephroureterectomy | | | May-10 |
| dhanusha | 4/f | 2008 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Dec-09 |
| wahithabanu | 4/f | 2008 | 3 | L | FH | triphasic | l.nephroureterectomy | | | Nov-09 |
| abimanu | 4/m | 13.06.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | Jun-09 |
| Anitha | 1y6m | 3.04.07 | 5 | b/l | FH | triphasic | | CT&RT | | lostf/u |
| barath | 10months | 4.12.07 | 1 | L | FH | triphasic | l.nephroureterectomy | | | Aug-09 |
| Darshini | 1y6m | 10.07.07 | 1 | R | FH | triphasic | r.nephroureterectomy | | | May-10 |
| dinesh | 10months | 30.04.07 | 1 | L | FH | triphasic | l.nephroureterectomy | | | Apr-09 |
| dathatreyan | 1y6m | 24.01.08 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| gouthami | 6/f | Jun-04 | 3 | R | FH | triphasic | r.nephroureterectomy | thrombus not extracted | | f/u-24.5.08 |
| ganesan | 1y6m | 26.06.06 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Sep-09 |
| gayathri | 2y6m | 11.07.06 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Oct-09 |
| harshavardan | 4/m | 10.06.06 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jan-10 |
| kamesh | 3y6m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jul-09 |
| kalaiselvan | 6/m | | 3 | L | FH | triphasic | | CT&RT | | lostf/u |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| monisha | 4y6m | | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| niranjan | | Mar-06 | 3 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Feb-09 |
| parthiban | 2y9m | 19.01.06 | 3 | L | FH | triphasic | l.nephroureterectomy | CT&RT | | Aug-09 |
| purosathaman | 5/m | 18.07.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | Apr-10 |
| praveen | 7/m | | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Mar-10 |
| pooja | 1/f | 19.09.06 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| pooja | 11months | 18.01.08 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| Ravikumar | 4/m | 26.10.06 | 1 | L | FH | triphasic | L.nephroureterectomy | | | 16.9.08 |
| tamilarasan | 1y6m | 30.12.06 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | |
| tangamani | 3/m | 13.06.07 | 2 | R | FH | triphasic | r.nephroureterectomy | local recurrence | | lostf/u |
| anish | 11months | 2008 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Apr-10 |
| pavithra | 2/f | 2008 | 1 | L | FH | triphasic | L.nephroureterectomy | cysticwilm | | May-10 |
| mohanraj | 3/m | 2008 | 2 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Sep-09 |
| sudharsana | 7/m | 1.7.03 | 3horse shoe kidney | | FH | triphasic | L.nephroureterectomy | tumour spill | | f/u-23/5/06 |
| pushpakar | 4y6m | 2003 | 2 | L | FH | triphasic | L.nephroureterectomy | CT&RT | | Jul-09 |
| pavithra | 2/f | 2003 | 2 | R | FH | triphasic | r.nephroureterectomy | CT&RT | | Jun-09 |
| parthiban | 1y6m | 2003 | 1 | L | FH | triphasic | l.nephroureterectomy | | | |
| syedsulthana | 5mon | 2003 | 1 | R | FH | triphasic | r.nephroureterectomy | | | |
| sheela | 4/f | 2003 | 2 | L | FH | triphasic | L.nephroureterectomy | ct&rt | | Feb-20 |
| sharmila | 3y6m | 2004 | 1 | R | FH | triphasic | r.nephroureterectomy | 522799 | 2008-well | |
| sundarrajan | 8/m | 2005 | 2 | R | UFH | triphasic | r.nephroureterectomy | | | lost f/u |
| surya | 5/m | 2005 | duplex-2 | R | FH | triphasic | r.heminephrectomy | 446864 | recurrence | defaultedRT |
| sakthivel | 10months | 2006 | 4 | L | FH | triphasic | L.nephroureterectomy | | | Oct-09 |
| Thirumalai | 4/m | 2006 | 2 | L | FH | triphasic | L.nephroureterectomy | | | Nov-09 |
| thirunavukarasu | 9/m | 2006 | 2 | L | FH | triphasic | L.nephroureterectomy | | | Jun-09 |
| thilani | 5/f | 2005 | 1 | L | FH | triphasic | L.nephroureterectomy | | | Sep-08 |
| thirumurugan | 5/m | 1.2.2006 | 3 | R | FH | triphasic | r.nephroureterectomy | 524468 | f/u9.9.08 | |
| vasanthkumar | 7/m | Mar-06 | | L | FH | triphasic | L.nephroureterectomy | 506170 | | Sep-09 |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| yuvaraj | 1y6m | Sep-06 | | L | FH | triphasic | L.nephroureterectomy | | | Jan-10 |
| yasmin | 4/f | Apr-06 | | L | FH | triphasic | L.nephroureterectomy | | | Apr-09 |
| hemadevi | 1y9m | 3.4.04 | 2 | R | FH | triphasic | r.nephroureterectomy | | | Feb-10 |
| lavanya | 8/f | 23.02.05 | 3 | R | FH | triphasic | r.nephroureterectomy | lung RT-givenf/u-15.7.08 | | |
| kaviarasan | | 1997 | | L | FH | triphasic | L.nephroureterectomy | | 2008-well | |
| sangeetha | | | 3 | L | FH | triphasic | L.nephroureterectomy | 8yr-survival | | |
| soubarnika | | 97 | | R | FH | triphasic | r.nephroureterectomy | 11yr-survival | | |
| sathishkumar | | | | R | FH | triphasic | r.nephroureterectomy | 15yr-survival | | |
| Binnsajan | 6/m | 568257 | 2 | L | FH | triphasic | l.nephroureterectomy | recurrence | lung sec | |
| kamurunisha- | 12/f | | 2 | L | FH | triphasic | l.nephroureterectomy | hemihypertrophy | | May-10 |
| muthumadhavan | 6/m | | duplex-3 | | FH | triphasic | l.nephroureterectomy | | | |
| thanusha | 4/f | | 3 | R | FH | triphasic | atrial thrombus-partial thrombectomy | | | |
| kumaran | 3y6m | 473179 | 3 | R | FH | triphasic | l.nephroureterectomy | thrombectomy | | f/u2009-feb |
| chaitanya | 3/f | | 3 | R | FH | triphasic | tumor dissemination | | neoadjuvani | |
| rohith | 2/m | | 3 | L | FH | triphasic | l.nephroureterectomy | biopsy | death | |
| malarvizhi | 5/f | 674021 | 1 | R | FH | triphasic | r.nephroureterectomy | hpe195/10 | | |
| deepika | | | 3 | L | FH | epithelial | l.nephroureterectomy | | neoadjuvant | |
| sudharsan | 1y6m | 672054 | | R | FH | triphasic | r.nephroureterectomy | | | |
| deepak | 2/m | 674525 | | l | FH | triphasic | l.nephroureterectomy | | | |
| sabarivasan- | 2/m | 673 | 2 | l | FH | triphasic | l.nephroureterectomy | | WAGR-syndrome | |
| monisha | 2/F | 674101 | 4 | R | FH | Rhabdomyomatous | r.nephroureterectomy | | neoadjuvant | |
| soundarya | 6/f | 673833 | 3 | R | FH | Rhabdomyomatous | r.nephroureterectomy | ascites | neoadjuvant | |
| aswathdevi | 1y6m | 676082 | 3 | R | FH | Rabdomyomatous | l.nephroureterectomy | | neoadjuvant | |

| Name | Age & Sex | F/U | Stage | | HPE | Hpe | Surgery | Chemo & RT | | Last F/U |
|---|---|---|---|---|---|---|---|---|---|---|
| jeganbabu | 4M | 664680 | | L | FH | | r.nephroureterectomy | | | |
| Harish | 3y6m | 658953 | 4 | R | FH | triphasic | r.nephroureterectomy | lung sec | death | |
| nivedha | 3/F | | 2 | L | FH | triphasic | l.nephroureterectomy | | | Apr-10 |
| Alifa | 9months | | 4 | L | UFH | rhabdoid | l.nephroureterectomy | liver sec | death | |
| senbagavel | 3/m | | 3 | R | FH | triphasic | | | neoadjuvant | |

# DISCUSSION

This is a retrospective study of 135 Wilms' tumor patients & prospective study of 21 patients from 2007 - 2010 . We have studied 99 male children &57 female children with Wilms tumor. We have studied 30 cases of stage-I disease ,61 cases of stage -II disease ,46 cases of stage -III disease ,10 cases of stage -IV Wilms , 9 cases of stage-V Wilms. We had followed NWTS -5 protocol.

All wilms tumor patient details were retrieved from tumor register & medical records department.

All children underwent follow-up screening with Ultra sonogram, CHEST X-ray , urine - micro albumin estimation , & blood pressure recordings . Plain x-ray of chest to look for secondary in lungs . CT chest was done in two cases which had doubtful shadows in chest x-rays.

## HISTOPATHOLOGY

Histopathology of 156 patients showed 2 unfavorable histology, 154 showed favorable histology[FH] ,which includes 145 triphasic histology & 9 monophasic variety

| Stage-I | Stage-II | Stage-III | Stage-IV | Stage-v |
|---|---|---|---|---|
| 30 | 61 | 46 | 10 | 9 |
| 27-triphasic<br>3-monophasic | 57-triphasic,<br>4-monophasic | 44-FH,2-unfavorable histology | 8-triphasic,<br>2-monophasic | 9-favorable histology |

Only 2 out of 156 cases were of unfavorable histology. Both cases were in stage III.

ULTRASONOGRAM was done to look for liver metastasis .

Presence of thrombus is picked up with high resolution ultrasound in IVC & atrium . They were serially screened for its disappearance with radiotherapy and chemotherapy**. It takes 24 weeks to two years for the tumor thrombus to resolve. We had six cases of tumor thrombus. 4 cases of IVC thrombus resolved with chemotherapy. One out of 2 atrial thrombus underwent**

**cardiac bypass and atrial thrombectomy . The other case of atrial thrombus resolved with chemotherapy**.

CT-contrast study was done in all bilateral tumor after chemo & RT to look for residual lesion.

5 year survival is 98% in stage 1 & 2. Long term disease free survival over 10 years is seen in 10 of our wilms tumor patients.

- Hypertension was present in 1% after ten years.
- We have had no second malignant neoplasm in our series.
- Micro albuminuria was present in1% of long term survivors.
- Their skeletal growth was affected in two children in our series .
- Ionising radiation affects epiphyseal growth.soft tissue hypoplasia and diminished bone growth is followed by scoliosis our series of patients had stunted growth but no scoliosis
- They did not show any delay in sexual maturity .
- Damage to the reproductive systems may represent one of the main late sequelae of both,gonadal radiation or chemotherapeutic agents.

- Radiation effects even on prepubertal germ cells may lead to gonadal dysfunction.

- Ovarian failure was seen in 75% of patients where the ovaries were included in radiation field in Europian study series.

- Ovaries are not included in radiation field in our centre. Hence we have not had ovarian failure in our series.

- Normal ovarian follicles were seen in Ultra-sonogram done in long term survivors in our centre.

- cardio toxicity due to adriamycin was not seen in our series.

ECHO was done in all stage III patients every year to rule out cardiotoxicity

- Renal failure was noted in 2 of our bilateral Wilms tumor cases who underwent bilateral partial nephrectomy.

- We have had radiation enteritis in 3 patients and

- one patient had defaulted RT and landed up with bilateral lung secondaries.

# OBSERVATION

- All bilateral wilms patients were under 7 years of age. Stages I & II have good prognosis. 97 % of cases show disease free survival status. Mortality is nil in our study from1998 till date in stages I &II .

- We have had 10 recurrences in stage III .Heminephrectomy was done in one of double moiety and child was not given radiotherapy as child was understaged Salvage chemotherapy was not effective in these children .

- Tumour spillage and defaulting chemotherapy or radiotherapy results in recurrences.In stage IV, 50% mortality was seen. Syndromic variety tumors do well with surgery, postoperative chemotherapy & radiotherapy[RT]. Bilateral Wilms have bad prognosis .Life expectancy in bilateral wilms' is two years from the time of onset of disease .

- Neo-adjuvant chemotherapy reduces recurrences &mortality in stage IV disease .Neo-adjuvant chemotherapy changes the histology of the tumour.

| Stage | Micro Albuminuria | Cardiac status | Skeletal growth | Sexual maturity | Hypertension |
|---|---|---|---|---|---|
| I & II | 2 | N | N | No delay | N |
| III | 15 | N | N | No delay | N |
| IV | RF: 2 | N | On F/U | On F/U | N |
| V | RF-2 | CCF-2 | Stunted-2 | - | N |

Foot-note: RF-renal failure, CCF-congestive cardiac failure,F/U-follow-up

Infertility:

Alkylating chemotherapeutic agents more often affect dividing cells and causing gonadal dysfunction. Vincristine has been described as a major risk factor for azoospermia and it is temporary. Our series children are too young to be checked.

# RESULTS

Prognosis in wilms tumor is directly dependant on the stage of disease .This emphasizes the need for accurate staging before chemotherapy & radiotherapy . Quality of life was well preserved in stages -1, 2 & 3 disease.

Recurrences are more common in tumor spillage and recurrent tumors have uniformly failed to respond to salvage regimen . Those children with advanced stage disease who responded favorably to chemo therapy & radiotherapy regime have survived beyond five years.The non-responders of this group deteriorated over the next five years.

We have had follow-up stage I & II children till 20 years of life & observed 97% disease free survival status .We have followed stage III children upto 10 years of their lives & observed 70% of disease free survival status .

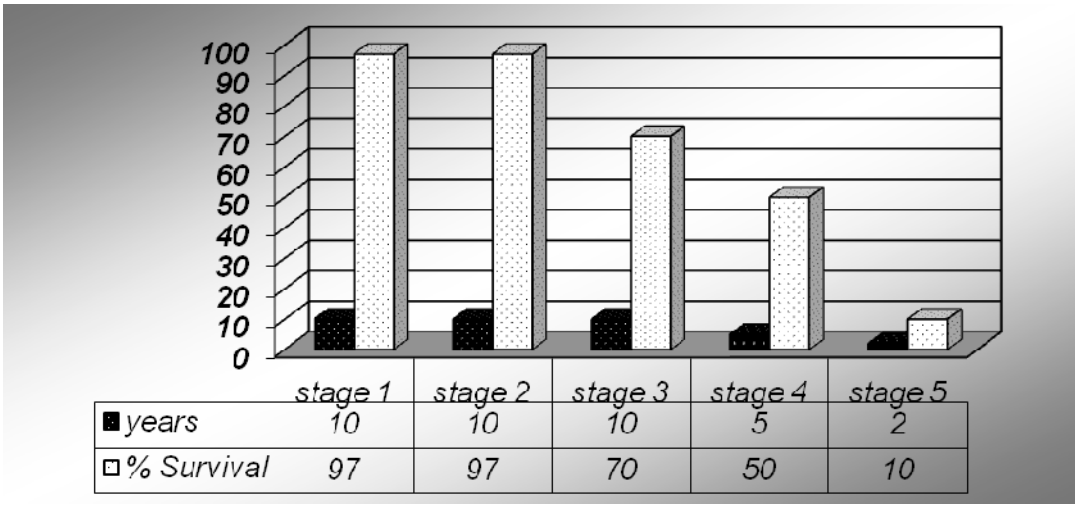
We have followed stage IV Wilms tumor children for 5 years & observed 50% mortality in our series .Stage V have shown 2 years survival from the onset of disease .

Stages I & II have good prognosis. 97 % of cases show disease free survival status. Mortality is nil in our study from1998 till date in stages I &II. We have had 10 recurrences in stage III. Salvage chemotherapy was not effective in these children.

In stage IV, 50% mortality was seen.

Syndromic variety tumors do well with surgery, postoperative chemotherapy & radiotherapy[RT]. Bilateral Wilms have bad prognosis .Life expectancy in bilateral wilms' is two years from the time of onset of disease .We had given neoadjuvant chemotherapy for 14 cases,9bilateral cases 5 large tumours,1ascites patient.

**Neo-adjuvant chemotherapy reduces recurrences &mortality in stage IV disease**.



# CONCLUSION

- Stage I &II have good prognosis .
- Stage III & IV needs close surveillance since they have high rate of recurrence. Recurrent tumors are not amenable to salvage chemotherapy.
- Stage V have bad prognosis.
- Stage IV wilms need lung irradiation.
- Neoadjuvant chemotherapy reduces tumor spillage in stage III &IV.
- Survival is poor in patients with wilms tumor and coexisting renal anomalies like horse shoe kidney

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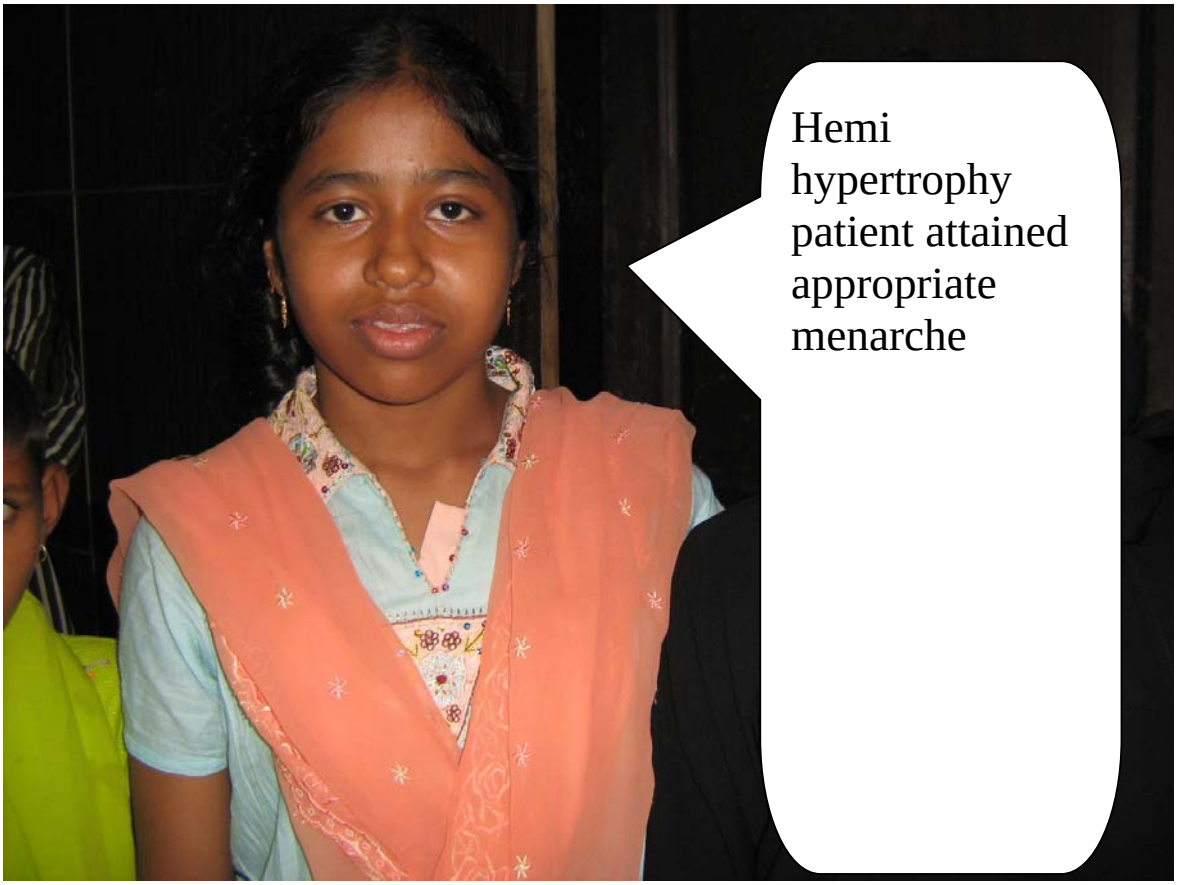
Hemi hypertrophy patient attained appropriate menarche